Quantitative Analysis of Preoperative Left Heart Volume in Tetralogy of Falot : The effect on the results and Hemodynamics after total repair

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Quantitative Analysis of Preoperative Left Heart Volume in Tetralogy of Fallot The effect on the results and hemodynamics after total repair

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Introduction

Remarkable improvement in surgical technique and intra- and post-operative management has decreased the mortality rate in cases of total repair for tetralogy of Fallot, even in infancy[2,3,6,25,26]. Nevertheless, contraindicative factors for primary repair remain. It has been reported that hypoplasia of peripheral pulmonary arteries results in a hypoplastic left heart which leads to postoperative left heart failure[10,12]. Severe hypoplastic left heart in cases of tetralogy of Fallot, despite a moderate size of the main pulmonary trunk, is a rare complication, however, such must not be overlooked by surgeons before total correction, because such patients inevitably die due to postoperative left heart failure with lung edema, even with an adequate relief of stenosis in the right ventricular outflow tract and a complete closure of the ventricular septal defect. Although some patients with hypoplastic peripheral pulmonary arteries may survive the total correction, postrepair assessment of hemodynamics may disclose a pulmonary hypertension[13].

The present study was a retrospective quantitative analysis of the preoperative left heart volume following death due to severe left heart failure and of survivors with postoperative pulmonary hypertension. The values were compared with those of the patients who had an excellent postoperative course and hemodynamics.

Material and Methods

One hundred and ninety-seven Japanese patients of tetralogy of Fallot were surgically

Key words: Hypoplastic peripheral pulmonary arteries, Hypoplasia of left heart, Left heart failure, Left atrial maximal volume, left ventricular end-diastolic volume, Postoperative pulmonary hypertension.
索引語：末梢肺動脈の発育不全，左心低形成，左心不全，左房最大容積，左室拡張末期容積，術後肺高血圧症.
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Table I. Age distribution and mortality in early and late series of total correction for tetralogy of Fallot.

Date of operation	Age at operation						Total	Mortality
	4–12mo.	1–2yr.	2–3yr.	3–4yr.	4–5yr.	5–31yr.		
Sept. 1968	16	17	15	12	10	18	88	16%
~ March 1973	(4)	(2)	(3)	(1)	(1)	(3)	(14)	
April 1973	—	10	28	37	12	22	109	7%
~ Dec. 1979		(1)	(1)	(5)	(0)	(1)	(8)	
Total	16	27	43	49	22	40	197	11%
	(4)	(3)	(4)	(6)	(1)	(4)	(22)	

( ): Death(s)

corrected over the eleven-year period from September 1968 to December 1979, in the Heart Institute of Kenritsu Amagasaki Hospital. The patients ranged in age from 4 months to 31 years. One hundred and twenty-nine cases were men and 68 were women. Of all patients, 80% were under five years of age (Table I). Twenty-two patients out of 197 (11%) were lost, but the results from April 1973 to December 1979 have remarkably improved (mortality rate 7%). Clinical investigation revealed that 12 patients died of right heart failure due to residual pulmonary stenosis, three from cerebral damage and 7 due to severe left heart failure (Table 2). Routine cardiac catheterization and angiocardiography were performed in the survivors about a month postoperatively. There were 18 with pulmonary hypertension; over 40 mmHg of pulmonary arterial systolic pressure, despite the lack of any residual cardiac anomalies[23].

It has been mentioned that decreased pulmonary blood flow as a result of a hypoplastic peripheral pulmonary artery makes the left heart very small and leads to postoperative pulmonary hypertension or severe left heart failure[10,12]. Thus, the left atrial maximal volume (LAMax) and left ventricular end-diastolic volume (LVEDV), on the preoperative angiocardiograms were assessed quantitatively. No remarkable bronchial collateral vessels were demonstrated in any of the angiocardiograms. LAMax was calculated excluding the volume of left atrial appendage. The left atrium is generally not well delineated when the pulmonary blood flow is greatly decreased, and in such cases, left atriography was necessary to obtain the left atrial volume. Here

Table II. Causes of death after total correction.

Causes	No. of patients
Right heart failure due to insufficient repair of right venticular outflow tract stenosis	12
Cerebral damage	3
Left heart failure	7

Table III. Preoperative left atrial and left ventricular volumes: patients died postoperatively of left heart failure (Group I).

Case No.	Age (Years)	BSA (m^2)	LAMax		LVEDV	
			cm^3/m^2	% Normal	cm^3/m^2	% Normal
1	2.3	0.48	8.4	25	31.1	59
2	4.3	0.51	8.6	24	32.5	60
3	4.6	0.59	13.2	36	31.6	55

Legend: BSA, body surface area. LAMax, left atrial maximal volume. LVEDV, left ventricular end-diastolic volume.

a catheter was passed from the right atrium through the patent foramen ovale.

Volume calculation was performed utilizing a special regression equation for infancy and childhood[11], that is, a modification derived from the biplane area-length method[7]. Only the informative preoperative angiocardiograms with a clearly delineated left atrium and/or left ventricle were used in the study and classifications were made, according to the postoperative results and hemodynamic states.

Group I: Three patients who died of severe left heart failure after correction (cases 1 to 3 in Table III).

Group II: 14 with postoperative pulmonary hypertension; greater than 40 mmHg of pulmonary arterial systolic pressure despite of without residual intracardiac anomalies (cases 4 to 17 in Table IV).

Table IV. Preoperative left atrial and left ventricular volumes: patients with postoperative pulmonary hypertension (Group II).

Case No.	Age (years)	BSA (m^2)	LAMax		LVEDV	
			cm^3/m^2	% Normal	cm^3/m^2	% Normal
4	0.9	0.38	—	—	78.1	163
5	0.9	0.41	9.5	29	—	—
6	1.0	0.39	13.3	42	62.0	128
7	1.0	0.38	—	—	44.7	93
8	1.3	0.43	15.6	46	83.4	197
9	1.9	0.44	10.9	32	40.7	80
10	2.8	0.45	9.3	27	58.7	114
11	3.2	0.53	—	—	38.5	70
12	3.3	0.61	9.4	25	—	—
13	4.1	0.60	—	—	51.8	89
14	4.6	0.57	11.2	31	—	—
15	4.7	0.75	14.2	36	43.4	68
16	7.8	0.82	8.9	25	—	—
17	10.3	1.01	17.5	39	67.7	93

Legend: BSA, body surface area. LAMax, left atrial maximal volume. LVEDV, left ventricular end-diastolic volume.

Table V. Preoperative left atrial and left ventricular volumes: patients with excellent postoperative hemodynamics (Group III).

Case No.	Age (years)	BSA (m^2)	LAMax		LVEDV	
			cm^3/m^2	% Normal	cm^3/m^2	% Normal
18	1.5	0.43	27.6	83	48.3	76
19	1.8	0.49	18.4	54	38.4	72
20	1.9	0.49	23.8	69	46.4	87
21	2.0	0.47	18.2	54	40.7	78
22	2.0	0.54	40.9	114	33.4	56
23	2.3	0.58	33.6	92	50.6	84
24	2.4	0.46	16.8	49	47.3	91
25	2.5	0.48	21.2	62	44.6	84
26	2.5	0.53	32.6	92	38.5	65
27	2.8	0.50	25.1	71	38.9	67
28	2.8	0.53	36.8	104	49.3	89
29	2.8	0.56	17.6	49	39.4	70
30	3.0	0.52	15.7	44	36.3	66
31	3.2	0.54	17.6	49	35.3	63
32	3.2	0.52	13.0	37	32.9	60
33	3.8	0.63	14.3	38	48.2	81
34	4.2	0.56	13.6	37	46.8	78
35	4.4	0.69	32.0	82	36.2	59
36	5.4	0.66	12.5	32	45.0	74
37	6.3	0.68	19.3	50	37.9	62

Legend: BSA, body surface area. LAMax, left atrial maximal volume. LVEDV, left ventricular end-diastolic volume.

Group III: 20 survivors after repair and with an uneventful postoperative course and excellent hemodynamics at postrepair cardiac catheterization; a right ventricular to aortic systolic pressure ratio less than 0.50 and without pulmonary hypertension (cases 18 to 37 in Table V).

All calculated volume values were divided by body surface area (m^2) in order to normalize data for patients with a different body surface area. Normal predicted LAMax was calculated from the following regression equation derived from the calculation data of LAMax in angiocardiograms of twenty-one without cardiac anomalies, in our institute;

$$\text{LAMax } (cm^3) = 43.9\ (\text{BSA})^{1.33}$$

normal LVEDV values were calculated from regression equation;[20]

$$\text{LVEDV } (cm) = 72.5\ (\text{BSA})^{1.43}$$

Each volume variable was also expressed as a percentage of normal by the equation;

$$\text{Volume value} = \frac{\text{calculated value}}{\text{normal predicted value}} \times 100 = \%\ \text{of normal}$$

Group comparisons were made using Student's t test to obtain the statistical values.

In addition, eight out of 18 patients with postoperative pulmonary hypertension underwent recatheterization from 14 months to 9 years after repair (mean 4.5 years).

Results

All data in each group are presented in Tables III, IV and V. Statistical analysis for these data is shown in Table VI.

LAMax values in group I were significantly ($p<0.01$) small, compared with those of group III and ranged from 8.4 to 13.2 cm^3/m^2, or 24 to 36% of normal. Fig. 1 shows a right ventriculogram of case No. 2 in which pulmonary arteries were hypoplastic and the small left atrium was insufficiently opacified. LAMax values in group II were also significantly ($p<0.001$) smaller than those of group III and averaged 12.0 ± 3.0 cm^3/m^2 (standard deviation), or $33\pm7\%$ of normal (Fig. 2).

As to LVEDV values, those in group I were significantly ($p<0.01$) smaller than those of group III. Only those in whom the LAMax and LVEDV values were both obtained are shown in Fig. 4. The values are represented as a percent of normal. The numbers in Fig. 4 correspond with the case numbers in Tables III, IV and V. Group III included three with smaller LVEDV values than those in group I; less than 60% of normal (case No. 22, 32 and 35). In all these patients, LAMax values were much larger than those in group I and averaged $78\pm39\%$ of the normal. Such probably explains why these patients did not succumb to critical postoperative left heart failure with lung edema. The postoperative course of case No. 9 was complicated.

Table VI. Statistical analysis of the data.

	LAMax			LVEDV		
	cm^3/m^2	% Normal	P (VS Group III)	cm^3/m^2	% Normal	P (VS Group III)
Group I (n= 3)	8.4—13.2	24— 36	<0.01	31.1—32.9	55— 60	<0.01
	10.1± 2.7	28± 7		31.9± 0.9	58± 3	
Group II (n=10)	8.9—17.5	25— 46	<0.001	38.5—83.4	68—197	<0.05
	12.0± 3.0	33± 7		56.9±15.8	110± 42	
Group III (n=20)	12.5—40.9	32—114		32.9—50.6	56— 91	
	22.5± 8.6	63± 24		41.7± 5.7	73± 11	

Legend: LAMax, left atrial maximal volume. LVEDV, left ventricular end-diastolic volume.

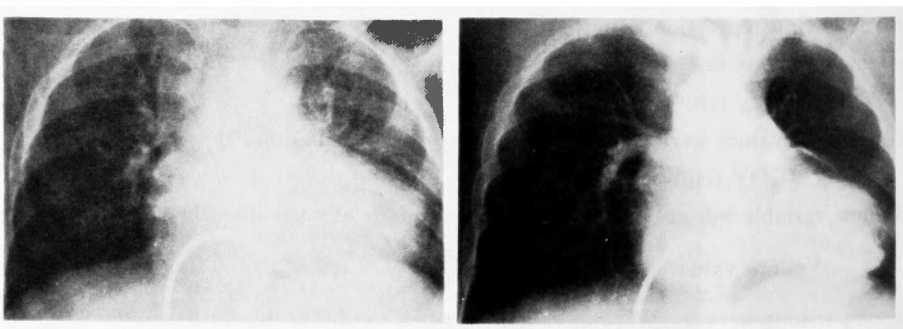

Fig. 1. Preoperative anteroposterior early (left) and late (right) stage views of injection into the right ventricle in case No. 2. Scattered peripheral pulmonary arteries are hypoplastic (left) and the left atrium is barely opaque in the late stage (right).

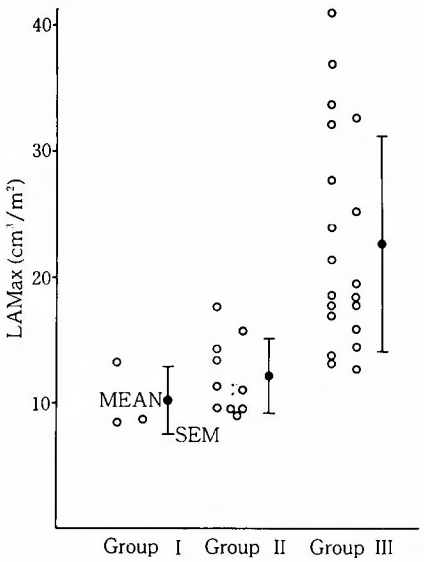

Fig. 2. The calculated left atrial maximal volume (LAMax) values are plotted for each of the three groups. The values in group I and group II are significantly smaller than those in group III ($P<0.01$ and 0 $P<0.001$, respectively).

Pulmonary edema became evident soon after she was weaned from cardiopulmonary bypass and bloody and foam-like aspiration continued for several hours. Mean arterial blood pressure, monitored from a radial artery, did not exceed 60 mmHg, despite a high left atrial pressure and administration of adequate doses of catecholamines. Although she barely survived the operation, severe low cardiac output persisted for two months after repair. Postoperative cardiac catheter-

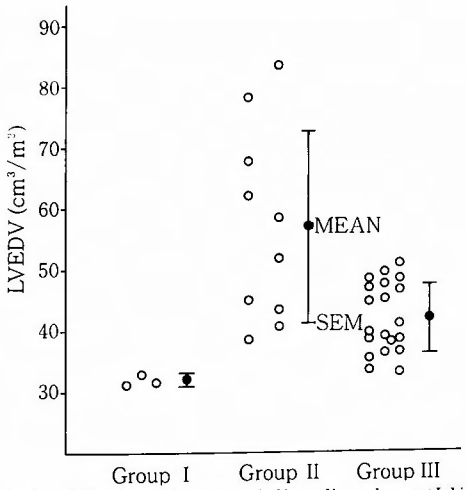

Fig. 3. The calculated left ventricular end-diastolic volume (LVEDV) values are plotted for each of the three groups. The values in group I are significantly ($P<0.01$) smaller than those in group III. Why the values in group II are significantly ($P<0.05$) greater than those in group III is uncertain (see text).

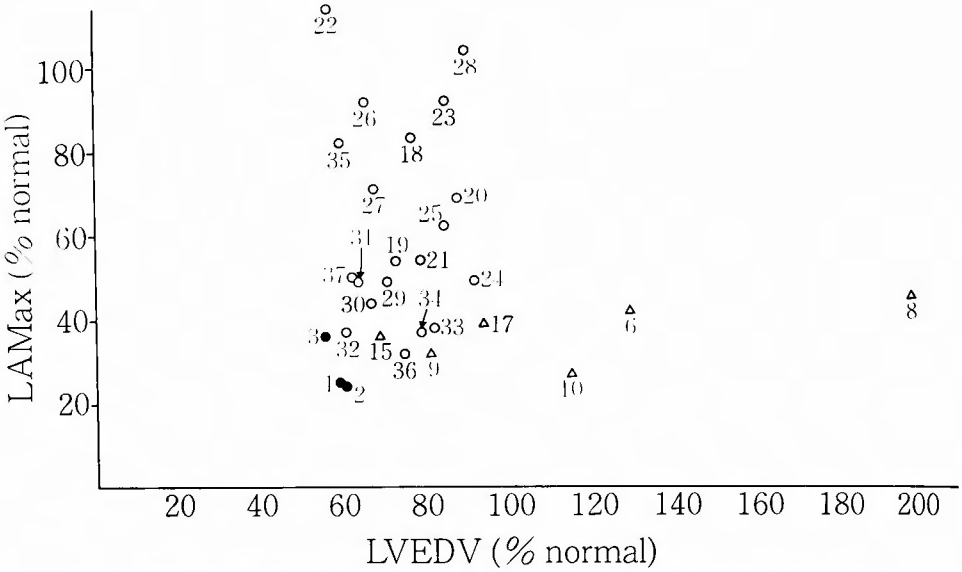

Fig. 4. Only those in whom left atrial maximal volume (LAMax) and left ventricular end-diastolic volume (LVEDV) are both available in Table III, IV and V are displayed. The numbers in this figure correspond with the case number in Table III, IV and V. Closed circles represent group I cases, triangles, group II cases and open circles, group III cases (see text).

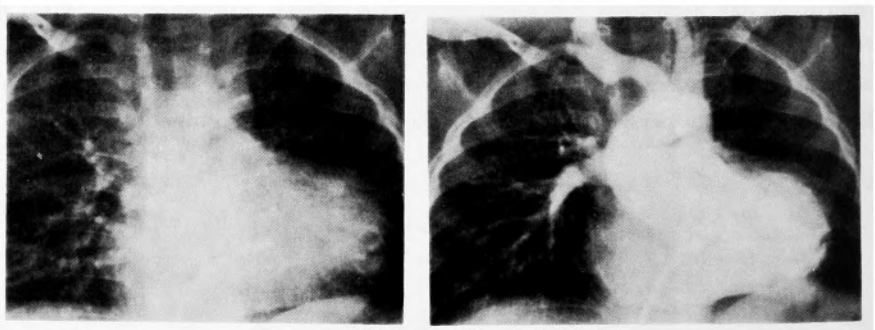

Fig. 5. Right ventriculograms of case No. 9 before total correction. Peripheral pulmonary arteries are hypoplastic, particularly on the left side despite the moderate size of main pulmonary artery (left). Small left atrium is not clearly demonstrated in the late stage (right). This patient survived total correction with a complicated postoperative course and pulmonary hypertension.

ization was done just before her discharge from hospital and a pulmonary hypertension without any residual ventricular septal defect was evident. Fig. 5 demonstrates underdeveloped peripheral pulmonary arteries, particularly on the left side, and small left atrium in case No. 9, before total repair.

Eight out of 18 patients, in whom the pulmonary hypertension was assessed by cardiac catheterization about one month after repair, underwent late recatheterization. These data are shown in Table VII. In the patients who had been treated at a relatively younger age (range 1

Table VII. Results of early and late catheterization studies on 18 patients with postoperative pulmonary hypertension.

Case No. in Table IV	Age at operation (yr.)	PASP (mmHg)		Interval from operation to recatheterization
		Early	Late	
4	4	50	—	
5	1	41	27	1 11/12 yr.
6	2	59	—	
7	2	41	—	
8	2	55	—	
9	3	41	—	
10	3	45	48	2 8/12 yr.
11	3	51	23	8 10/12 yr.
12	3	54	—	
13	4	42	27	1 2/12 yr.
14	4	75	—	
—	4	53	36	1 2/12 yr.
15	5	51	—	
—	5	56	37	9 1/12 yr.
16	7	43	—	
17	10	48	46	2 4/12 yr.
—	13	53	45	9 yr.
—	31	45	—	

Legend: PASP, pulmonary arterial systolic pressure.

to 5 years old), pulmonary arterial systolic pressure appreciably decreased from the early levels, except for one patient, three years of age at the time of correction (case No. 10). Two patients who had undergone total correction at an older age (10 and 13 years old) had high levels of pulmonary arterial systolic pressure; 45 and 46 mmHg, respectively.

Discussion

The remarkable improvements in surgical technique have resulted in decreases in the rate of mortality in cases of total correction of tetralogy of Fallot. The surgical techniques in total repair of tetralogy of Fallot include (1) complete closure of ventricular septal defect (2) adequate enlargement of the stenotic right ventricular outflow tract and (3) closure of a patent ductus arteriosus and a patent foramen ovale[16], should such exist. Details of the surgical techniques used for the enlargement of the outflow tract stenosis are now well established[14,19,21,22].

Although most pediatric cardiologists carefully observe the degree of severity of right ventricular outflow tract stenosis in tetralogy of Fallot, cardiac surgeons must also examine the left heart when they discuss the indications for complete repair. It has been reported that tetralogy of Fallot may be accompanied with hypoplastic peripheral pulmonary arteries[8], small left atrium[5,24] and small left ventricle[15,17,18].

Remarkable hypoplasia of peripheral pulmonary arteries and decreased pulmonary blood

flow make the left ventricle so small that total correction is not feasible[10,12]. The incidence of hypoplastic left heart may be rare[18], nevertheless, cardiac surgeons should investigate its possibility before attempting correction.

Quantitative analysis of left ventricle as an indicative factor for primary repair of tetralogy of Fallot was first described by JARMAKANI et al.[12] They reported that the patient with only 60% of normal LVEDV value died of left heart failure after total repair. GRAHAM and COLLEAGUES[10] also mentioned that only those with a 65% or greater of the normal LVEDV value should be considered for primary repair and in those with 55% or less of normal values, shunt operation should be done as a first procedure. They also stated that values between 55 and 65% might be an indication for an initial shunt procedure, but sufficient data to support this had not been obtained.

From the data obtained in the present study, the degree of development of left heart as an indication for primary repair cannot be simply assessed only by LVEDV, but the LAMax should also be calculated. Our data suggest that those with a small LVEDV (less than 60% of normal) and also a small LAMax (less than 40% of normal) should not be considered for primary repair, because postoperative left heart failure and lung edema occur in most cases. Even when LVEDV values are small but over 56% of the normal, primary repair can be successfully performed, unless the LAMax is less than 40%. Patients with a remarkably small value of LVEDV, for example less than 45% of normal, as described by GRAHAM et al.[10], may not be good candidates for primary correction even though the size of LAMax is moderate.

Children with a small LAMax and normal LVEDV can survive the primary repair but postoperative pulmonary hypertension will ensue, as presented in Table IV. Why the LVEDV in group II is significantly ($p<0.05$) larger than those in group III (Fig. III) is not clear. Such may be due to the large blood flow from the right to the left ventricle, through the ventricular septal defect, with simultaneous direct flow from the right ventricle to the aorta, as described COELHO et al.[5] The approximately normal size of the left ventricular volume in Group II was considered to prevent severe left heart failure postoperatively, despite a small LAMax, but did consequently lead to postrepair pulmonary hypertension. Postoperative pulmonary hypertension is an unfavorable hemodynamic state and improvements did occur at recatheterization, in the late period, only among younger children. This implies that obstructive vascular changes in peripheral pulmonary arteries will occur in older children who have undergone no surgical intervention[1], or that neogenesis of the peripheral pulmonary vessels may be already impaired in adolescence.

To increase the volume of the left heart, shunt procedure is effective[12], however, patch enlargement of the right ventricular outflow tract, without closure of the ventricular septal defect, may be effective for those with a very small calibre of right or left pulmonary artery. This provides for the benefit of symmetrical pulmonary artery growth[9,26].

Summary and conclusions

Preoperative left heart volume of tetralogy of Fallot was quantitatively investigated as one

of the indicative factors for primary repair. Left atrial maximal volume (LAMax) and left ventricular end-diastolic volume (LVEDV) on preoperative angiocardiogram were calculated in three patients who had died of left heart failure and lung edema, in 14 survivors associated with postoperative pulmonary hypertension and also in 20 patients with an uneventful postoperative course and excellent hemodynamics at the time of cardiac catheterization after repair. All volume values were obtained by Dodge's biplane area-length method and were corrected by Graham's regression equations for infants and children. The values were divided by the individual's body surface area and were also expressed as a percent of the normal. The conclusions are as follows:

(1) In cases where the LAMax is over 40% of the normal, primary repair is indicated if the LVEDV is over 60% of normal.

(2) In cases where the LAMax is $33 \pm 7\%$ of the normal and the LVEDV is within a normal range, the child can survive the total correction, but postoperative pulmonary hypertension does occur. In such cases, total repair should not be postponed, as pulmonary hypertension remains in the late period if surgery is done during adolescence.

(3) Those with a very small LAMax (less than 40% of normal) and also with a very small LVEDV (less than 60% of normal) should undergo a primary shunt procedure or mere relief of right ventricular outflow tract stenosis without closure of ventricular septal defect, with subsequent total correction after the left heart grows enough.

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和文抄録

左心低形成を伴うファロー四徴症根治手術の臨床的研究：とくに左房・左室容積の根治手術成績ならびに術後血行動態に及ぼす影響に関する検討

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ファロー四徴症に対する手術手技の進歩に伴い，その根治手術成績は乳幼児においてさえ近年めざましく向上しつつある．今後さらに死亡率の低下を図るための一手段として，その手術適応を厳密に決定する必要がある．

ファロー四徴症のなかには，末梢肺動脈の発育不全がきわめて高度で肺血流量が少ないため，左房・左室の低形成を伴う症例があり，この左心低形成が著しいものは根治手術後に肺水腫を伴う重篤な左心不全を呈して死亡する．したがって，術前における左心発育度の綿密な評価は根治手術成績向上を図る上で重要である．

昭和54年12月までの約11年間に兵庫県立尼崎病院心臓センターで施行されたファロー四徴症根治手術症例は197例であるが，そのうち7例を左心不全で失った．また，生存しえても術後心カテーテル検査で心室中隔欠損遺残や末梢肺動脈狭窄を認めないにもかかわらず肺高血圧症を呈した症例を18例経験した．そこで，左心不全による死亡例7例のうち，術前の心血管造影で左房または左室像の明瞭なもの3例（Ⅰ群），同じく根治手術後肺高血圧症を呈した18例のうちから14例（Ⅱ群）を選び，術後血行動態良好な20症例（Ⅲ群）を対照として左房最大容積（LAMax）および左室拡張末期容積（LVEDV）を算出したところ次のような結果を得た．

A．Ⅰ群ではLAMax，LVEDVともに小さくLAMaxは正常値の28±7%，LVEDVは58±3%であった．

B．Ⅱ群ではLAMaxはⅠ群同様小さいにもかかわらず（33±7%），LVEDVはほぼ正常値に近かった（110±42%）．

C．Ⅲ群20例のLVEDVは正常値の73±11%であったが，56～60%の症例が3例存在した．しかし，これらの症例のLAMaxはいずれも比較的大きく，正常値の78±39%であった．

D．術後肺高血圧症を呈した18例のうち8例に遠隔期心カテーテル検査を施行したところ，根治手術時年令5歳以下の6例中5例には肺高血圧症の改善がみられたが，根治手術時年齢10歳および13歳の2例では遠隔期にもなお肺高血圧症の残存を認めた．

以上の結果よりファロー四徴症左心容積の根治手術成績および術後血行動態に及ぼす影響について以下の結論を得た．

1．ファロー四徴症根治手術適応としての左心容積を評価するに当っては，従来のようにLVEDVの測定のみでなく，LAMaxの測定もあわせ行うのが妥当と考えられる．

2．LAMaxが正常値の40%以上，かつLVEDVが正常値の60%以上あれば根治手術に耐えうる．

3．LVEDVが正常値に近くてもLAMaxが40%以下の症例では，根治手術後生存しえても術後肺高血圧症を呈する．このような症例では学童期以前に根治手術を施行しておけば，遠隔期での肺高血圧症の改善を望みうる．

4．LAMaxが正常値の40%未満かつLVEDVが正常値の60%以下の症例では，まず姑息手術を施行し左心の発育を促した後での二期的根治手術が望ましい．